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Year: 2018

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## **Progressive functional underdrainage in CSF shunt dependent women during pregnancy. Case report and review of the literature**

Krauss, Philipp ; Fritz-Naville, Marco ; Regli, Luca ; Stieglitz, Lennart Henning

DOI: <https://doi.org/10.1016/j.wneu.2017.09.108>

Posted at the Zurich Open Repository and Archive, University of Zurich

ZORA URL: <https://doi.org/10.5167/uzh-141922>

Journal Article

Accepted Version



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Originally published at:

Krauss, Philipp; Fritz-Naville, Marco; Regli, Luca; Stieglitz, Lennart Henning (2018). Progressive functional underdrainage in CSF shunt dependent women during pregnancy. Case report and review of the literature. *World Neurosurgery*, 109:372-376.

DOI: <https://doi.org/10.1016/j.wneu.2017.09.108>

## TITLE PAGE

**Title:**

Progressive functional underdrainage in CSF shunt dependent women during pregnancy. Case report and review of the literature

Authors:

(corresponding Author)

**1. Ph. Krauss / Philipp, KRAUSS**

Department of Neurosurgery, University Hospital of Zurich,  
Frauenklinikstrasse 10, 8091 Zurich, Switzerland

[Philipp.Krauss@usz.ch](mailto:Philipp.Krauss@usz.ch), Phone: 0041 (0) 44 255 1111

**2. M. Fritz / Marco FRITZ-NAVILLE, MD**

Gynecologist, Zeltweg 4 8032 Zurich, Switzerland

**3. L. Regli / Luca REGLI, MD**

Department of Neurosurgery, University Hospital of Zurich,  
Frauenklinikstrasse 10, 8091 Zurich, Switzerland

**4. L. H. Stieglitz / Lennart Henning STIEGLITZ, MD**

Department of Neurosurgery, University Hospital of Zurich,  
Frauenklinikstrasse 10, 8091 Zurich, Switzerland

Key words: hydrocephalus, ventriculoperitoneal shunt, pregnancy, intraabdominal pressure, intracranial pressure

Word count (without figures and tables): 1224; Figures: 2; Tables: 1

\*Disclosure-Conflict of Interest [authors to provide own statement, .doc(x) format preferred]

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**Conflict of interest statement:**

The authors declare that the article content was composed in the absence of any commercial or financial relationships that could mean a potential conflict of interest.

**Abbreviations:**

CSF - (cerebrospinal fluid), VP-Shunt - (ventriculoperitoneal shunt), SVS - (slit-ventricle syndrome), VA-Shunt - (ventriculoatrial shunt), LP-Shunt - (lumboperitoneal shunt), VPL-Shunt - (ventriculopleural shunt), com - (communicating); non com - (non communicating), He - (headache), na - (nausea), vom - (vomiting), vag - (vaginal), ces - (cesarean), PP shunt - (perfusion pressure shunt), ICP - (intracranial pressure), HP - (hydrostatic pressure), CP - (closing pressure valve), IAP - (intraabdominal pressure)

## **INTRODUCTION:**

Due to advances in the treatment of hydrocephalic patients since the 1960s, women suffering from hydrocephalus nowadays may live normal lives including sexuality and pregnancy. Thus, obstetricians and neurosurgeons encounter pregnant women with cerebrospinal fluid (CSF) diversion devices more often. Still, there are often questions among physicians regarding management during pregnancy. Early symptoms of elevated intracranial pressure (ICP), such as headache, nausea and vomiting regularly occur during the course of pregnancy. A precise differentiation between shunt dysfunction and pregnancy-related symptoms is important to minimize risk to mother and child due to ICP exacerbation or unnecessary surgical shunt revisions. Some cases of shunt dysfunctions during pregnancy are reported in the literature, still their natural history remains unclear. We report the case of a 34 years-old secundipara with ventriculoperitoneal shunt (VP-shunt) who showed progressive need of elevated CSF drainage during pregnancy, returning to pre-pregnancy status just after delivery along with a pubmed review of the English literature. To our knowledge, this is the only reported case of conservative treatment by consecutive change of valve pressure level correlating pregnancy progression and valve pressure.

## **CASE REPORT:**

A 34 years-old woman para 1, gravida 2, was followed at the department of neurosurgery at the university hospital of Zurich, Switzerland. Early in her childhood she was diagnosed with a congenital communicating hydrocephalus and treated with a VP-shunt. She underwent several shunt revisions hence. Her condition was complicated by a slit-ventricle syndrome, as it is common among patients treated with shunt systems not actively countering the siphoning effect during childhood and adolescence. The current implanted system consists of an intra- and extracranial Bactiseal catheter (Codman/DepuySynthes, New Brunswick, USA) in the right frontal horn of the lateral ventricle and a ProGAV 2.0 (Miethke, Potsdam, Germany) shunt valve with a ShuntAssistant 0-25cmH<sub>2</sub>O (Miethke). The Patient's condition was stable at a valve

pressure level of 10cmH<sub>2</sub>O without signs of CSF over- or underdrainage. At the 18th gestational week of her second pregnancy she was admitted suffering from headache suggesting CSF underdrainage. Clinical and laboratory examination showed no signs of mechanical shunt obstruction, disconnection or inflammation. Valve pressure level was reduced from 10cmH<sub>2</sub>O to 9cmH<sub>2</sub>O which resulted in immediate symptom regression the following day.

During the course of her pregnancy, the patient returned several times with signs of underdrainage. Every time, quick symptom relief was achieved by subsequently adjusting the valve pressure level from 10cmH<sub>2</sub>O to 5cmH<sub>2</sub>O. No obstetrical complications occurred. The patient was followed by her obstetrician and underwent gynecological exams and ultrasonography regularly. In the 39th gestational week the patient delivered a healthy infant through planned cesarean section. Immediately after delivery, the patient developed positional headache and diplopia suggesting CSF overdrainage. Again, symptom relief was quickly achieved by raising the valve pressure level to 8cmH<sub>2</sub>O, then 9cmH<sub>2</sub>O several days later and finally stabilized asymptotically at 10cmH<sub>2</sub>O about 13 weeks after delivery (Figures #, ##).

## **DISCUSSION:**

### Overview of published cases:

Pregnancy in women with CSF diversion devices remains a rare constellation, still. Consequently, our knowledge is mostly based on case reports and several case series. In 1979 the first two cases of hydrocephalic women observed during pregnancy were reported.<sup>1</sup> The largest published case series retrospectively analyzed 138 pregnancies in 70 shunt-dependent women.<sup>2</sup> Conclusions were drawn from a detailed auto-questionnaire. To date 117 shunt dependent women with 204 pregnancies are reported in the English literature (see Table).

### Risk of delivery in shunt dependent women:

Miscarriages were reported in 31 cases reflecting 15.2% of all pregnancies with a clustering of 13 miscarriages in only three women. Five therapeutic abortions were

performed of whom four were because of hydrocephalus.<sup>2,3</sup> In 18 cases cesarean section was chosen because of the hydrocephalic condition of the mother.<sup>2,4-7</sup> Postpartal VP-shunt infection only occurred in one patient.<sup>8</sup> All other reported cases showed the birth of healthy infants without complications regardless the technique of delivery.

#### Shunt malfunction during pregnancy:

The incidence of shunt malfunction during pregnancy varies widely in the literature from no signs of malfunction<sup>6,9-13</sup> up to 59% of pregnancies with signs of elevated ICP.<sup>14</sup> Liakos et al. describe a shunt revision rate of approximately 20% during pregnancy.<sup>2</sup> Symptoms range from transient headache, nausea, vomiting<sup>1,2,4-6,14-19</sup> and visual disturbances<sup>5,6,20</sup> to severe persistent neurological deficits<sup>7</sup> or miscarriage.<sup>3</sup>

#### Treatment of shunt malfunction during pregnancy:

In case of shunt malfunction, different treatment options are reported. Cesarean section,<sup>2,4-7</sup> transforming VP-shunts into VA-shunts,<sup>18-21</sup> endoscopic ventriculostomy as alternative treatment for symptomatic occlusive hydrocephalus,<sup>22,23</sup> conservative management by recurrently aspirating CSF from the valve reservoir<sup>16</sup> or repeated flushing of the pumping device are described with good clinical results.<sup>7,15,17</sup>

#### Physiology of CSF diversion through VP-shunt during pregnancy:

To date, the physiology of pregnancy related shunt malfunction remains unclear. The obstruction of the peritoneal shunt catheter between intraabdominal organs<sup>15</sup> or a change in the pressure gradient between valve and intraabdominal pressure (IAP) are proposed explanations.<sup>16,20</sup> Distal cavity pressure is involved in the pressure gradient leading to CSF drainage<sup>(+)</sup><sup>24,25</sup> and raised IAP can result in a higher shunt threshold.<sup>26</sup> In these conditions a reduced overall resistance of the shunt system is needed to provide physiological CSF flow. In patients with fixed pressure gravitational units its opening pressure is chosen due to body height mainly. Thus, gravitational valves may lead to underdrainage in selected patients and especially during gravidity. Programmable gravitational units or flow-control devices could address this problem. Still, a linear prediction of compartmental pressure seems little accurate using this equation in fully implanted systems.<sup>27</sup>

$$PPshunt = (ICP + HP) - (CP + IAP)$$

(+) PP shunt = perfusion pressure shunt; ICP = intracranial pressure; HP = hydrostatic pressure; CP = closing pressure valve; IAP = intraabdominal pressure

Chronically raised IAP correlates with BMI and abdominal diameter<sup>28</sup> and intraperitoneal fluid or constipation may result in VP-shunt malfunction<sup>29,30</sup>. However, short intraabdominal hypertension seemingly does not lead to shunt malfunction.<sup>31</sup> During pregnancy the IAP is chronically raised and quickly falls to regular levels after delivery.<sup>32-34</sup> Likewise, short peaks of IAP<sup>35,36</sup> do not seem to affect the ICP as vaginal delivery can be performed without complications.<sup>1,2,6,9,10,12,14-16,18-20,22,23,37</sup>

#### Future perspectives:

Only few years ago adjustable shunt valves were commonly used for older patients mainly and the slit- ventricle syndrome (SVS) was a common phenomenon in patients with infantile hydrocephalus. In these patients raised IAP during pregnancy may counteract a preexisting chronic overdrainage and even result in temporary symptom relief. Today, a generation of women, treated with adjustable valves, reaches an age where reproduction is possible. In these patients drainage equilibrium can be altered during pregnancy and result in shunt malfunction. Therefore, shunt malfunction during pregnancy may become a more frequent problem, especially in cases where gravitational valves are used. As typical symptoms of shunt malfunction such as headache, nausea or vomiting regularly occur during pregnancy, shunt malfunctions may remain undetected in many cases. This can lead from unnecessary discomfort up to serious health risk. We propose that pregnant shunt-dependent women should be closely followed by obstetricians and neurosurgeons together to detect cases of shunt malfunction early. In these cases a cranial MRI can be helpful to verify the malfunction and we propose valve pressure level adjustment as conservative and safe treatment option.

**CONCLUSION:** In CSF shunt dependent women, pregnancy-related changes in the CSF drainage equilibrium can occur and become symptomatic. CSF underdrainage can quickly be managed by adjustment of the valve pressure level, avoiding invasive procedures. Physicians should be vigilant to discriminate hydrocephalic from pregnancy related



symptoms. In young female patients adjustable gravitational units could facilitate the conservative management during pregnancy. Therefore the implantation of such devices should be considered in young women who might possibly plan pregnancy in the future.

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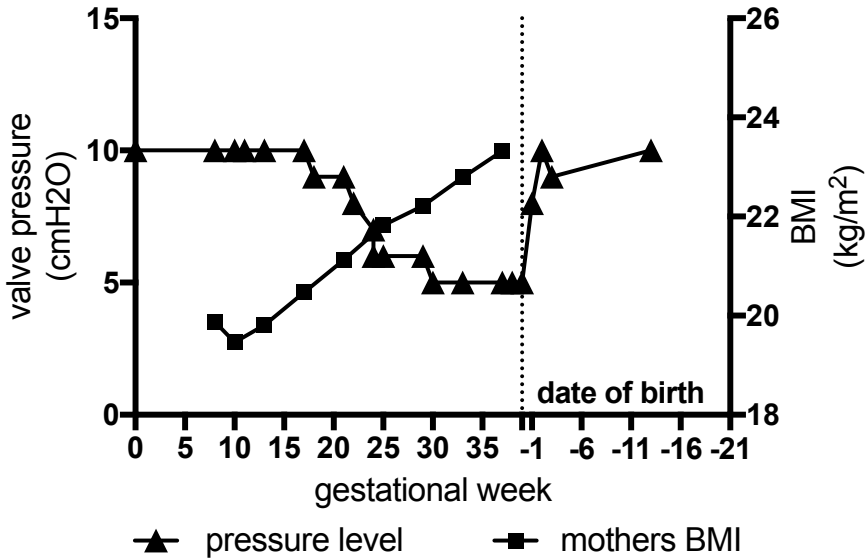
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Table(s)  
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	Patients/ Pregnancies	Type of CSF Shunt	Type of Hydrocephalus	Shunt complications	related treatment	delivery
Monfared et al. 1979	2	VP Shunt	non com.	"blackouts"	none	vag.
		no prior shunt	non com.	He., na., vom.	VA Shunt Placement	vag.
Howard&Herrick 1981	2	VP Shunt	com.	none	none	vag.
		no prior shunt	non com.	memory loss, paresthesia	VP Shunt Placement	vag.
Kleinman et al. 1983	2	VP Shunt	com.	He., na., vom., gaze palsy	pumping shunt	vag.
		VP Shunt	non specified	He., na., vom.	pumping shunt	vag.
Gast et al. 1983	2/3	VA Shunt	non com.	imp. vigilance, ataxia	none	vag.
Hanakita et al. 1985	1	VP Shunt	non com.	He., blurred vision, imp. vigilance	VP to VA Shunt transformation	vag.
Fröhlich et al. 1986	3	VP Shunt	non com.	none	none	ces.
		VP Shunt	non specified	none	none	vag.
		no prior shunt	non specified	none	none	vag.
		VA Shunt	com.	none	none	vag.
Hassan & Moumani 1988	2	2 VP Shunt		none	none	
Samuels et al. 1988	2	VP Shunt	non com.	imp. vigilance	preterm cesarean	ces.
		VP Shunt	com.	He., na., vom., gazy palsy	preterm cesarean	ces.
Houston & Clein 1989	1	VP Shunt	non specified	He.,na.,vom.	CSF aspiration	vag.
Wisoff et al. 1991	18/21, 10/11 original cases	16 VP Shunt, 3 VA Shunt, 1 VPL Shunt	non specified	10 he., 6 na./vom., 5 imp. vigilance, 3 ataxia, 3 gaze palsy	4 Shunt Revisions in original cases	3 vag. 8 ces.
Okagaki et al. 1990	1	VP Shunt			VP to VA Shunt transformation	
Cusimano et al. 1990	1	VP Shunt	non com.	He., na., dizziness	CSF aspiration and pumping	ces.
Olatunbosun et al 1992	1	VP Shunt	non com.	none	none	vag.
Landwehr et al. 1994	8/25	4 LP Shunt, 4 VP Shunt	4x IIH, 4x non specified	none	none	14 vag. 4 ces. 7 abort.
Kurtsoy et al. 1994	1	no prior shunt	non com.	Na., vom., ataxia	VP Shunt then VA Shunt Placement	vag.
Goolsby & Harlass 1996	1	VP Shunt	non specified	Imp. vigilance, diabetes insipidus	pumping shunt	ces.
Liakos et al. 2000	70/138	54/84 VP Shunt, 10/16 VA Shunt, 1/1 VPL Shunt, 1/2 LP Shunt, 5/5 Mult. Configuration	various (for detailed listing see liakos et al. 2000)	8 he., 15 transient symptoms of raised ICP	Revision prior to delivery 7, Revisions following delivery 23, Revision folloqing miscarriage	61 vag., 44 ces., 30 Misc.
Kane et al. 2003	1	VP Shunt	com.	post partum shunt infection	VP Shunt replacement	ces.
Fletcher et al. 2007	2	VP Shunt	non com.	none	none	vag.
		VP Shunt	non specified	He., na., vom., incontinence	preterm cesarean	ces.
Yoshida et al. 2007	1	VP Shunt	non com.	Imp. vigilance, dizziness	VP Shunt to EVT Conversion	vag.
Rees et al. 2008	1	VP Shunt	non specified	status epilepticus	VP Shunt revision	misc.
Murakami et al. 2010	1	VP Shunt	com.	He., na., vom., gaze palsy	VP Shunt to VA Shunt conversion	vag.
Schiza et al. 2012	1	VP Shunt	non com.	He., na., vom., gaze palsy, dysphasia	preterm cesarean	ces.
total patiens/pregnancies	117/204	95 VP Shunt, 15 VA Shunt, 5 LP Shunt, 2 VPL Shunt, 4 no prior shunt, 5 mult. Configurations				

#)

Valve pressure vs. mothers BMI



##)

### Valve pressure vs. fetal weight

